```
          RAW SEQUENCE LISTING              DATE: 11/06/2000
          PATENT APPLICATION: US/09/639,207   TIME: 11:21:18

          Input Set : A:\Sequence Listing - 06618-686001.txt
          Output Set: N:\CRF3\11062000\I639207.raw

   4  <110> APPLICANT: Kazemi-Esfarjani, Parsa
   5        Benzer, Seymour
   7  <120> TITLE OF INVENTION: AN ANIMAL MODEL OF POLYGLUTAMINE
   8        TOXICITY, METHODS OF USE, AND MODULATORS OF POLYGLUTAMINE
   9        TOXICITY
  11  <130> FILE REFERENCE: 06618-686001
  13  <140> CURRENT APPLICATION NUMBER: US 09/639,207
  14  <141> CURRENT FILING DATE: 2000-08-14
  16  <150> PRIOR APPLICATION NUMBER: US 60/148,934
  17  <151> PRIOR FILING DATE: 1999-08-12
  19  <150> PRIOR APPLICATION NUMBER: US 60/148,933
  20  <151> PRIOR FILING DATE: 1999-08-12
  22  <150> PRIOR APPLICATION NUMBER: US 60/177,047
  23  <151> PRIOR FILING DATE: 2000-01-18
  25  <150> PRIOR APPLICATION NUMBER: US 60/205,720
  26  <151> PRIOR FILING DATE: 2000-05-19
  28  <160> NUMBER OF SEQ ID NOS: 69
  30  <170> SOFTWARE: FastSEQ for Windows Version 4.0
  32  <210> SEQ ID NO: 1
  33  <211> LENGTH: 508
  34  <212> TYPE: PRT
  35  <213> ORGANISM: Human
  37  <400> SEQUENCE: 1
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  39   1               5                   10                  15
  40  Ser Ser Asn Ser Glu Met Asp Val Glu Ile Thr Thr Glu Gln Pro Thr
  41               20                  25                  30
  42  Ile Asp Val Lys Ala Glu Gln Ile Val Pro Lys Asp Ala Ala Thr Ile
  43               35                  40                  45
  44  Ala Glu Glu Lys Lys Lys Leu Gly Asn Asp Gln Tyr Lys Ala Gln Asn
  45           50                  55                  60
  46  Tyr Gln Asn Ala Leu Lys Leu Tyr Thr Asp Ala Ile Ser Leu Cys Pro
  47  65                  70                  75                  80
  48  Asp Ser Ala Ala Tyr Tyr Gly Asn Arg Ala Ala Cys Tyr Met Met Leu
  49               85                  90                  95
  50  Leu Asn Tyr Asn Ser Ala Leu Thr Asp Ala Arg His Ala Ile Arg Ile
  51               100                 105                 110
  52  Asp Pro Gly Phe Glu Lys Ala Tyr Val Arg Val Ala Lys Cys Cys Leu
  53           115                 120                 125
  54  Ala Leu Gly Asp Ile Ile Gly Thr Glu Gln Ala Val Lys Met Val Asn
  55           130                 135                 140
  56  Glu Leu Asn Ser Leu Ser Thr Ala Val Ala Ala Glu Gln Thr Ala Ala
  57  145                 150                 155                 160
  58  Gln Lys Leu Arg Gln Leu Glu Ala Thr Ile Gln Ala Asn Tyr Asp Thr
  59               165                 170                 175
  60  Lys Ser Tyr Arg Asn Val Val Phe Tyr Leu Asp Ser Ala Leu Lys Leu
  61               180                 185                 190
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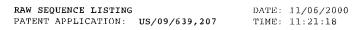
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 63         195             200             205
 64 Phe Leu Gly Arg Cys Asp Glu Ala Leu Asp Ile Ala Val Ser Val Met
 65     210             215             220
 66 Lys Leu Asp Thr Thr Ser Ala Asp Ala Ile Tyr Val Arg Gly Leu Cys
 67 225             230             235             240
 68 Leu Tyr Tyr Thr Asp Asn Leu Asp Lys Gly Ile Leu His Phe Glu Arg
 69             245             250             255
 70 Ala Leu Thr Leu Asp Pro Asp His Tyr Lys Ser Lys Gln Met Arg Ser
 71         260             265             270
 72 Lys Cys Lys Gln Leu Lys Glu Met Lys Glu Asn Gly Asn Met Leu Phe
 73         275             280             285
 74 Lys Ser Gly Arg Tyr Arg Glu Ala His Val Ile Tyr Thr Asp Ala Leu
 75     290             295             300
 76 Lys Ile Asp Glu His Asn Lys Asp Ile Asn Ser Lys Leu Leu Tyr Asn
 77 305             310             315             320
 78 Arg Ala Leu Val Asn Thr Arg Ile Gly Asn Leu Arg Glu Ala Val Ala
 79             325             330             335
 80 Asp Cys Asn Arg Val Leu Glu Leu Asn Ser Gln Tyr Leu Lys Ala Leu
 81         340             345             350
 82 Leu Leu Arg Ala Arg Cys Tyr Asn Asp Leu Glu Lys Phe Glu Glu Ser
 83         355             360             365
 84 Val Ala Asp Tyr Glu Thr Ala Leu Gln Leu Glu Lys Thr Pro Glu Ile
 85     370             375             380
 86 Lys Arg Met Leu Arg Glu Ala Lys Phe Ala Leu Lys Lys Ser Lys Arg
 87 385             390             395             400
 88 Lys Asp Tyr Tyr Lys Ile Leu Gly Ile Gly Arg Asn Ala Ser Asp Asp
 89             405             410             415
 90 Glu Ile Lys Lys Ala Tyr Arg Lys Lys Ala Leu Val His His Pro Asp
 91         420             425             430
 92 Arg His Ala Asn Ser Ser Ala Glu Glu Arg Lys Glu Glu Glu Leu Lys
 93         435             440             445
 94 Phe Lys Glu Val Gly Glu Ala Tyr Ala Ile Leu Ser Asp Ala His Lys
 95     450             455             460
 96 Lys Ser Arg Tyr Asp Ser Gly Gln Asp Ile Glu Glu Gln Glu Gln Ala
 97 465             470             475             480
 98 Asp Phe Asp Pro Asn Gln Met Phe Arg Thr Phe Phe Gln Phe Asn Gly
 99             485             490             495
100 Gly Gly Arg Asn Asn Ser Ser Phe Asn Phe Glu Phe
101         500             505
103 <210> SEQ ID NO: 2
104 <211> LENGTH: 2239
105 <212> TYPE: DNA
106 <213> ORGANISM: Drosophila
108 <400> SEQUENCE: 2
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110 acaaacaca aaattcaagt ttaaaaacta ataggcaac taaaagggaa gccgcagcga    120
111 ataaagtgat ttgctgaaag agacgtaaga aagttaatcg catcgaaggc accagaaatc    180
112 ggggatttct aacacggcgc gcgtgcgacg tacatacata cgcaagcgca cacacacacg    240
```

```
113 aacaattact tgccattgac gcaaaagcga aaaagcagtg gaataaaggg gaattgacaa      300
114 ataacaacgt tttgcaagca ctggactctg gtcgctggtg ttctttcatt ttgtaattgc      360
115 cacgcatgga cgacgaagta attgaaatta gcgacagcga acgcgaagaa acctcatcga      420
116 actccgaaat ggatgtggaa ataacgacag aacagccaac catcgatgtc aaagcagagc      480
117 aaattgtgcc caaggacgcg gcaaccattg ccgaggagaa gaagaaactg ggcaacgacc      540
118 aatacaaggc gcagaactat cagaatgcac tcaagctcta cacggatgcc atatcgctgt      600
119 gtccggactc ggcggcatac tatggcaatc gggccgcctg ctacatgatg ctgctcaact      660
120 ataatagcgc cctgaccgac gcccgacacg ccatacgcat cgatccgggc ttcgagaagg      720
121 cctacgtccg tgtggccaag tgctgtctgg ccctgggcga cattattggc accgaacagg      780
122 ccgtcaaaat ggtcaacgag ctgaattcgc ttagcacggc tgttgctgcc gaacagacgg      840
123 cggcgcaaaa gttgcgccaa ttggaggcca ccattcaggc gaactacgat acgaaatcct      900
124 atcgcaatgt ggtcttctat ttggatagtg ccttgaaatt ggcgccccgcc tgtttgaaat      960
125 atcgtctact caaggctgag tgccttgcat ttttggggcg atgtgatgag gccttggaca     1020
126 ttgcggtcag tgtaatgaaa ctggatacca catcggcgga tgcgatatac gtgagaggtc     1080
127 tgtgcctgta ctacacggac aacctggaca agggaattct tcatttcgag cgcgccctga     1140
128 ccctcgaccc ggaccactac aagtccaagc agatgcgcag caaatgcaag cagctcaagg     1200
129 agatgaagga gaacggcaat atgctattca agtcgggtcg gtatcgcgag gcacacgtta     1260
130 tctacacgga cgccctgaag atcgatgaac acaacaagga tatcaattcg aaattgcttt     1320
131 acaatcgggc tttggtcaac acgcgtattg gcaatttgcg agaggccgtg gccgattgca     1380
132 atcgagtgct ggagctgaat agtcagtatc tgaaggctct gttgctgcga gcgcgctgct     1440
133 acaatgatct ggagaagttc gaggagtcgg tggcggacta tgagacggcg ctgcagctgg     1500
134 agaagacgcc ggagattaag cgaatgctgc gcgacgccaa gtttgcgttg aagaagtcaa     1560
135 agcgaaagga ctactacaag atcctgggca ttggacgcaa tgcgtccgac gacgagatca     1620
136 agaaggcgta tcgcaaaaag gcgctggtac atcatccgga tcgacacgca aacagcagtg     1680
137 ccgaggagcg caaggaggag gagctcaagt tcaaggaggt gggcgagcg tacgccatac     1740
138 tgtcggatgc tcacaagaag tcgcgctacg acagcggcca ggatatcgag gagcaggagc     1800
139 aagccgactt cgatccgaat caaatgttcc gcacattctt ccaattcaac ggcggtggcc     1860
140 ggaataattc atcgttcaac tttgagttct aggatcccaa cgagtgttgt tcaccaccac     1920
141 agagaagaag accatctcaa tcccatactt tctgcctcat ccgaaaccaa catacagcag     1980
142 cgcacaaatt ttgaactctt ttacatattt cttttccaaa aagcaagaaa ataccacatt     2040
143 ttgattatgt taacgaatga atatatgcca agttatttga aaaaatattc taaatcaaaa     2100
144 taatgcaact aaatttccag tgtaagttca cattttttaaa tgttctttct tggatttttt     2160
145 tttcggcaac attaataaat catgggagat ttgtgttaaa taaacagaaa tatacatata     2220
146 aaaaaaaaaa aaaaaaaaa                                                   2239
148 <210> SEQ ID NO: 3
149 <211> LENGTH: 273
150 <212> TYPE: PRT
151 <213> ORGANISM: Drosophila
153 <400> SEQUENCE: 3
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155  1               5                  10                  15
156 Leu Met Asn Asn His Met Asn His Thr Met Asn Ala Met Asn Met Gln
157             20                  25                  30
158 Met Arg Ser Met Asn Arg Leu Met Asn Ser Phe Met Pro Asp Pro Phe
159         35                  40                  45
160 Met Gln Val Ser Pro Phe Asp Gln Gly Phe Gln Gln Asn Ala Leu Met
161     50                  55                  60
162 Glu Arg Pro Gln Met Pro Ala Met Pro Ala Met Gly Leu Phe Gly Met
163 65                  70                  75                  80
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RAW SEQUENCE LISTING
PATENT APPLICATION: US/09/639,207

DATE: 11/06/2000
TIME: 11:21:18

Input Set : A:\Sequence Listing - 06618-686001.txt
Output Set: N:\CRF3\11062000\I639207.raw

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164 Pro Met Met Pro Asn Phe Asn Arg Leu Leu Asn Ala Asp Ile Gly Gly
165             85              90              95
166 Asn Ser Gly Ala Ser Phe Cys Gln Ser Thr Val Met Thr Met Ser Ser
167             100             105             110
168 Gly Pro Asp Gly Arg Pro Gln Ile Tyr Gln Ala Ser Thr Ser Thr Lys
169             115             120             125
170 Thr Gly Pro Gly Gly Val Arg Glu Thr Arg Arg Thr Val Gln Asp Ser
171     130             135             140
172 Arg Thr Gly Val Lys Lys Met Ala Ile Gly His His Ile Gly Glu Arg
173 145             150             155             160
174 Ala His Ile Ile Glu Lys Glu Gln Asp Met Arg Ser Gly Gln Leu Glu
175             165             170             175
176 Glu Arg Gln Glu Phe Ile Asn Leu Glu Glu Gly Glu Ala Glu Gln Phe
177             180             185             190
178 Asp Arg Glu Phe Thr Ser Arg Ala Ser Arg Gly Ala Val Gln Ser Arg
179     195             200             205
180 His His Ala Gly Gly Met Gln Ala Ile Met Pro Ala Arg Pro Ala Ala
181     210             215             220
182 His Thr Ser Thr Leu Thr Ile Glu Pro Val Glu Asp Asp Asp Asp Asp
183 225             230             235             240
184 Asp Asp Asp Cys Val Ile Gln Glu Gln Gln Pro Val Arg Ser Ser Ala
185             245             250             255
186 Gly Arg His Tyr Ser Ser Ala Pro Thr Ala Pro Gln Asn Arg Tyr Asn
187             260             265             270
188 Tyr
191 <210> SEQ ID NO: 4
192 <211> LENGTH: 1753
193 <212> TYPE: DNA
194 <213> ORGANISM: Drosophila
196 <400> SEQUENCE: 4
197 ggcacgagga aaatattcgt gaaaattctg catacggaaa gaagaaaatt cgagcaacag        60
198 aaagccaaca caatccacaa aaatgtcttt attcggagcg ttgatgggtg atttcgacga       120
199 cgatctcggc cttatgaaca accacatgaa ccacactatg aacgcgatga acatgcagat       180
200 gcgctcgatg aatcgcctga tgaacagctt tatgcccgat cccttcatgc aggtctcgcc       240
201 ctttgaccag ggattccagc agaacgctct catggagcgt ccgcagatgc cggccatgcc       300
202 agccatgggc ctcttcggca tgccatgat gccaaacttt aatcgcctgt tgaacgctga       360
203 tattggtggc aattcaggcg catccttctg ccagagcacc gtgatgacca tgtcatcggg       420
204 tcccgatggg cgtcctcaga tctaccaggc cagcactagt accaaaaacag gaccgggagg       480
205 cgttcgtgag acccgcagga cggtgcagga ctcgcgcact ggggtgaaga agatggccat       540
206 tggtcatcac atcggcgagc gggcacacat tattgagaaa gagcaggaca tgcgctcagg       600
207 acaactggag gagcgccagg agttcattaa tctggaggag ggagaagccg agcagtttga       660
208 cagggagttt acatcgcgcg ctagtcgcgg agcggtgcag tcaagacatc atgctggtgg       720
209 catgcaggcc atcatgcccg cccgtccagc ggcacacacc tcgacgttga ccattgagcc       780
210 agtggaggac gacgacgacg atgatgatga ctgtgtaatc caggagcagc aaccggttcg       840
211 ctcctccgcg ggccgccatt attccagtgc gccaacggca ccgcagaaca gatataatta       900
212 ctaaatctaa agtcaataca gtatatttta ctaactatcc gataaaacag aaacagaatt       960
213 gcatactata aatttctgct aattacattc ccaactgcgt tcaaacgaaa cgaatatcga      1020
214 atcgaaatca tagaatgcac agagcagcat acatccacat ccctatgccg ccaatccgag      1080
215 gcgccaacaa cgtgccgtaa aacatttca cacggaggac gaagcggcca gctcctacaa      1140
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216 ggcggtcaag cgcggcaaga agaagtagta gaaacgtgat catctgtatg ccaacatctt   1200
217 ccgcatcgca cactcaaaaa cactaggaag caaagcgttg ggttctgttc catagcagga   1260
218 aaaccaattc aaatattttt taacaaacac aattctttac cagttctgtc ttatcctgcg   1320
219 tgagtcgacc agaatgcaac actaaaaaat gtacaacttc aagatgctat tgatgtgcac   1380
220 gcaggataca gaacaacttg cttaaattta cttaaaacaa atgtgactat tcaacgccga   1440
221 aatcattaca acacacactc tcagacctaa tcgaaaaatt caatgaaagt aatggaatat   1500
222 atatgaaatc gtaattataa gtttgaatta tttgattaat tctcaagttt ttagattttg   1560
223 ttagccacta agctttaaat tatggatgcc agttagcgtg caaatgaaca caattgattt   1620
224 gaaggctccg aacgatagaa aacaacaatt accaattccc caaatacatg taattcgtaa   1680
225 ggcctaagta aatgttaacg tgaatttaat taaatggtaa ttacattata atagtaaaaa   1740
226 aaaaaaaaaa aaa                                                      1753
228 <210> SEQ ID NO: 5
229 <211> LENGTH: 484
230 <212> TYPE: PRT
231 <213> ORGANISM: Human
233 <400> SEQUENCE: 5
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235  1               5                  10                  15
236 Glu Ala Glu Thr Phe Lys Glu Gln Gly Asn Ala Tyr Tyr Ala Lys Lys
237             20                  25                  30
238 Asp Tyr Asn Glu Ala Tyr Asn Tyr Tyr Thr Lys Ala Ile Asp Met Cys
239         35                  40                  45
240 Pro Lys Asn Ala Ser Tyr Tyr Gly Asn Arg Ala Ala Thr Leu Met Met
241     50                  55                  60
242 Leu Gly Arg Phe Arg Glu Ala Leu Gly Asp Ala Gln Gln Ser Val Arg
243 65                  70                  75                  80
244 Leu Asp Asp Ser Phe Val Arg Gly His Leu Arg Glu Gly Lys Cys His
245                 85                  90                  95
246 Leu Ser Leu Gly Asn Ala Met Ala Ala Cys Arg Ser Phe Gln Arg Ala
247             100                 105                 110
248 Leu Glu Leu Asp His Lys Asn Ala Gln Ala Gln Glu Phe Lys Asn
249             115                 120                 125
250 Ala Asn Ala Val Met Glu Tyr Glu Lys Ile Ala Glu Thr Asp Phe Glu
251     130                 135                 140
252 Lys Arg Asp Phe Arg Lys Val Val Phe Cys Met Asp Arg Ala Leu Glu
253 145                 150                 155                 160
254 Phe Ala Pro Ala Cys His Arg Phe Lys Ile Leu Lys Ala Glu Cys Leu
255                 165                 170                 175
256 Ala Met Leu Gly Arg Tyr Pro Glu Ala Gln Ser Val Ala Ser Asp Ile
257             180                 185                 190
258 Leu Arg Met Asp Ser Thr Asn Ala Asp Ala Leu Tyr Val Arg Gly Leu
259             195                 200                 205
260 Cys Leu Tyr Tyr Glu Asp Cys Ile Glu Lys Ala Val Gln Phe Phe Val
261     210                 215                 220
262 Gln Ala Leu Arg Met Ala Pro Asp His Glu Lys Ala Cys Ile Ala Cys
263 225                 230                 235                 240
264 Arg Asn Ala Lys Ala Leu Lys Ala Lys Lys Glu Asp Gly Asn Lys Ala
265                 245                 250                 255
266 Phe Lys Glu Gly Asn Tyr Lys Leu Ala Tyr Glu Leu Tyr Thr Glu Ala
```

Please Note:
**Use of n and/or Xaa have been detected in the Sequence Listing. Please review the
Sequence Listing to ensure that a corresponding explanation is presented in the <220> to
<223> fields of each sequence which presents at least one n or Xaa.**

VERIFICATION SUMMARY   DATE: 11/06/2000
PATENT APPLICATION: US/09/639,207   TIME: 11:21:19

Input Set : A:\Sequence Listing - 06618-686001.txt
Output Set: N:\CRF3\11062000\I639207.raw

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L:1278 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:16
L:1298 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:17
L:1338 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:19
L:1374 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:21
L:1429 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:24
L:1430 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:24
L:1455 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:25
L:1535 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:30
L:1540 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:30
L:1626 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:35
L:1649 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:36
L:1686 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:38
L:1761 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:42
L:1783 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:43
L:1840 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:46
L:1912 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:51
L:1917 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:51
L:1941 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:52
L:1961 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:53
L:2029 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:56
L:2140 M:341 W: (46) "n" or "Xaa" used, for SEQ ID#:62
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